Section of Physical Medicine

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DISCUSSION ON THE CLINICAL AND ELECTROMYOGRAPHIC ASPECTS OF POLYMYOSITIS

Professor F. J. Nattrass:

Polymyositis: Clinical Aspects

Among the very varied cases of flaccid paralysis it is necessary to recognize a group in which weakness and muscular wasting are due to diffuse inflammatory changes in muscles. Though a syndrome of polymyositis has been known and described by many authors for nearly a hundred years, renewed interest in it is warranted for two reasons:

(1) The frequent association of such muscular disease with skin changes—dermatomyositis being no longer a very uncommon diagnosis—and the recognition of the affinity of these cases with others of the so-called collagen-vascular or connective tissue diseases, especially scleroderma and lupus erythematosus.

(2) The recognition that polymyositis may occur with no skin changes, or with minimal skin changes, and that the clinical picture then closely resembles that of primary muscular dystrophy. Though such cases are uncommon, they are not so rare as to be unimportant, for the very practical reasons that the prognosis and treatment may be quite different from those of true muscular dystrophy. They present a fairly well-defined clinical picture and a histological picture in a muscle biopsy which is usually distinctive. They have a varied course, sometimes rapid in onset and progress, sometimes insidious and chronic; and they may occur either in childhood or in adult life. They can be separated clearly from septic or parasitic infections of muscles, e.g. trichiniasis, but their aetiology is as obscure as that of collagen-vascular diseases generally.

In trying to define the clinical pictures I shall deal first with a group of cases in childhood which are probably of this nature.

With the co-operation of Dr. John N. Walton I have recently studied, and published an account of, 8 children who recovered from an illness diagnosed in each case as muscular dystrophy (Nattrass, 1954). I shall mention some details of one or two only and summarize the others.

Case I.—This boy was normal till the age of 3 when he began to waddle from side to side and his abdomen stuck out when he walked. He tended to stumble and fall very easily and once on the floor he was unable to get up again by himself. Whereas previously he could climb stairs he now had to pull himself up by holding the banisters with his arms. Within a month of the onset of these symptoms his calves began to swell, particularly the right one which ached a good deal. I saw him at this stage and made the diagnosis of muscular dystrophy. He showed wasting of the pectorals, serrati, infra- and supra-spinati, and looseness of the shoulder girdles. There was marked lordosis with characteristic "climbing up the legs", the thigh muscles were weak and those of the calves were hypertrophied. Within a year the condition had progressed so much that he was unable to walk more than 100 yards by himself and had to be carried to school. The condition remained relatively stationary for another three months and then began to improve.

Constant encouragement on the part of the parents was a feature of his management. At the age of 6 he was able to run races at school. At the age of 11 he lived a perfectly normal life including football and gym, though it was perhaps true that he could not run as quickly as many boys of his own age. Examined in 1952, he was a healthy-looking boy who walked and ran perfectly well and rose from the floor without difficulty: in a word, nothing abnormal was found on detailed examination.

There can be no doubt of the diagnosis of muscular disease. However, it is very uncommon for myopathy to progress so rapidly that the patient is practically unable to walk within a year of the onset, and this relatively acute course is the most important atypical feature.

Case II.—This boy was perfectly well in the first three years of life. At the age of 3 he developed a disinclination to play with boys of his own age because he was unable to keep up with them. He was inclined to play on the floor and when he picked up a cup he used both hands, or put one hand under the other elbow to lift an object. Within three months he was unable to walk more than 50 yards, could not get up from the floor without assistance and was unable to climb a step 6 in. high. He then showed a waddling gait, excessive lumbar lordosis, weakness and wasting of shoulder girdle muscles, and pseudohypertrophy of the hamstring and calf muscles.

FEBRUARY
This boy was given a vitamin-B preparation for six months and later vitamin-E capsules. Improvement was observed within two to three weeks of beginning the first of these remedies and continued gradually but steadily. It was about eighteen months before he could walk upstairs easily and rise from the floor normally. In time he recovered virtually completely, to lead a normal active life, with only a trace of residual muscular wasting. As in the first case, the rapidity of the onset was, in retrospect, the feature most unlike true muscular dystrophy.

Of the remaining 6 cases, 3 appeared to skilled observers to be the subjects of pseudo-hypertrophic muscular dystrophy. The other 3 were in various respects less typical, though the diagnosis of dystrophy was made in all. In retrospect 2 may have been suffering from a form of benign congenital myopathy. 3 patients began to improve so soon after beginning treatment with vitamin-E products that it seems likely that this treatment influenced the disease. In the other patients recovery was probably spontaneous. In all recovery was virtually complete. There is no proof from muscle biopsy that these 8 patients, whose histories have been traced, and most of whom have been examined by us only since their recovery, were the subjects of polymyositis. Except, however, in 2 of the patients, this diagnosis is the most probable on the analogy of other cases seen subsequently in an acute phase of a similar illness. Certainly they were not the subjects of true muscular dystrophy.

I have seen several cases in adults which have been shown by biopsy to be suffering from polymyositis: in these there is a general similarity in the clinical picture, but with some important differences from the childhood cases.

Patient (Fig. 1) was seen at the age of 50 with a three-year history of increasing loss of power in the arms and shoulders. On examination, there was symmetrical wasting of the scapular and upper arm muscles and bilateral wasting and weakness of the face. There was slight weakness of the spinal and pelvic girdle muscles. He was thought to be a case of facioscapulohumeral dystrophy but with some very unusual features. There was no family history of any similar condition: slight wasting of the sternomastoids and hands was observed but there was little weakness of these. Above all the history was very short and the progress of the condition much more rapid than in this most benign form of muscular dystrophy: deterioration was indeed becoming serious (Fig. 1). An electromyogram showed changes consistent with a primary muscle disease and a muscle biopsy from the deltoid showed a florid picture of a subacute myositis (Fig. 2). Dr. Walton will deal with the histological changes and the relation to other connective tissue diseases.
This patient improved at once on cortisone. While much of the improvement in his appearance was attributable to the general effects of the hormone, there was also marked improvement in muscular power. This proceeded slowly for several months on a maintenance dose of 75 mg. daily, but at the present time, twelve months later, improvement seems to have stopped and muscular weakness is still severe.

I am left with the impression that adult cases of polymyositis resemble the facioscapulohumeral type or the limb-girdle (i.e. Erb) type of muscular dystrophy rather than the Duchenne or pseudohypertrophic type of childhood dystrophy. They differ from these forms of primary muscular dystrophy in that they may show involvement also of the muscles of deglutition, of the neck muscles, and to some extent of the hand muscles. A hint is sometimes given of the link with collagen-vascular diseases by a history of vascular phenomena of the Raynaud type. A clinical point which I cannot explain is that until an advanced stage, unlike muscular dystrophy, the tendon reflexes may be not only retained but unduly active.

Diagnosis is not rendered easier by the fact that true muscular dystrophy may appear first in adult life, but nevertheless it is likely that more and more cases so diagnosed may prove to be suffering from polymyositis. This conclusion almost certainly applies to the cases described by Shy and McEachern (1951) as “menopausal muscular dystrophy”, in which great improvement followed cortisone or wheat-germ oil therapy. It may well apply also to many other cases of apparently typical muscular dystrophy in early life which have been reported as greatly improved after treatment with wheat-germ and/or vitamin E or its analogues. There is very strong evidence that such treatment is entirely without effect in true muscular dystrophy (Walton and Nattrass, 1954).

Conclusions.—Polymyositis without involvement of skin or blood vessels occurring in childhood has, on the whole, a good prognosis and may be influenced by treatment.

Polymyositis in later life has a less favourable prognosis, but seems divisible into two groups:

1) Resembling a chronic muscular dystrophy and remaining stationary or deteriorating slowly over many years.

2) Associated with dermatitis and then often a much more severe illness, of shorter duration and high mortality.

Important aid in diagnosis from neuropathic disorders is given by electromyography: Dr. Bauwens will deal with the extent to which this investigation can differentiate polymyositis and muscular dystrophy.

I would by no means advocate even so relatively harmless a procedure as muscle biopsy in cases of typical muscular dystrophy with a family history. But when there are unusual features, such as a negative family history and especially either an unexpectedly rapid onset and course or the occurrence of remissions, then this aid to diagnosis is essential. In acute or subacute cases the histological picture is usually unmistakable; in the chronic cases the changes are more difficult to identify, but are generally distinctive unless destruction of muscle is very advanced.

REFERENCES


Dr. John N. Walton:

Polymyositis: Diagnosis, Pathology, Prognosis and Treatment

Although progressive muscular dystrophy has been recognized for almost a century as the classical example of a myopathic disorder, we have more recently seen descriptions in the literature of a large number of inflammatory, degenerative and metabolic disorders of muscle, some of which are apparently new. Among these we may mention dermatomyositis, polymyositis, neuromyositis, interstitial myositis in association with collagen disease, generalized myositis fibrosa, calcinosis universalis, menopausal muscular dystrophy and carcinomatous myopathy. Each of these conditions has been described in some detail, but in many instances the number of reported cases is few, and for this reason they are not well known. As Professor Nattrass has pointed out, this lack of general recognition is clearly shown by the fact that cases of polymyositis continue to be diagnosed as progressive muscular dystrophy. Moreover, definition of each of these conditions is often so inexact

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that one cannot determine whether we are dealing with a number of distinct diseases or simply with several variants of a few basic disorders of muscle.

It was with the object of reviewing critically some of these problems that I collected together, in collaboration with Dr. Raymond D. Adams of Boston, a series of 40 cases of what we have chosen to call "polymyositis". Clinically, all of the cases showed, at some stage of the illness, unmistakable symptoms and signs of muscle disease, with weakness and/or atrophy. Pathologically, all 34 cases in which muscle sections were obtained revealed a degenerative and/or inflammatory process unlike that of progressive muscular dystrophy. These observations have led us to conclude that the many conditions enumerated above are variants of a single clinical and pathological syndrome, namely, polymyositis.

We do not use the term polymyositis to imply that the disease is infective, or indeed that it is always inflammatory in nature; in certain cases there is undoubtedly evidence of an inflammatory reaction in the affected muscles, but in others which may be clinically identical such signs are absent. It may seem difficult to justify our terminology in describing cases of the latter type, but the terms dermatomyositis and polymyositis are now so well established that it would be difficult to change them; furthermore, it would not be satisfactory to call the condition "idiopathic polymyopathy", as this description would be equally apt for cases of muscular dystrophy. We feel, therefore, that the syndrome is best referred to as polymyositis while admitting that the term may be a misnomer in certain cases which show no inflammatory change. It is, of course, reasonable to continue to use the title dermatomyositis when there are skin changes or to qualify the diagnosis as, say, polymyositis with rheumatoid arthritis, when there are associated signs of a collagen disease.

Though some workers use the term polymyositis simply to identify a pathological picture, the pathological changes in muscle in the disease under discussion, though generally different from those of muscular dystrophy, are not distinctive, but may be seen in experimental vitamin-E deficiency, virus infections, poisoning with certain toxic agents and even, in very minor degree, following long confinement in bed. It is the combination of the characteristic clinical and pathological features which identify the syndrome of polymyositis, and we believe that the term should be utilized only to indicate this syndrome. The pathological change alone may properly be called a "myositis".

**CASES**

We have divided our cases arbitrarily into 4 clinical groups which are:

**Group I.**
- Polymyositis.
- Late life ("menopausal muscular dystrophy").
- Acute, with myoglobinuria.

**Group II.**
- Polymyositis with muscle weakness the predominant feature but with associated features of a collagen disease
- Dermatomyositis with predominant muscular weakness and with minimal or transient skin changes.

**Group III.**
- Collagen disease with muscular disability of secondary importance
- Dermatomyositis with florid skin changes and less obtrusive muscular involvement.

**Group IV.**
- Polymyositis ("carcinomatous myopathy") or dermatomyositis in association with malignant disease.

I do not mean to imply that each group is a clearly-defined nosological entity; but the groupings are useful for clinical purposes. During the course of a single illness one may see cases move from one group to another. Indeed, we had one patient who suffered episodes variously diagnosed as acute rheumatism, lupus erythematosus, dermatomyositis, calcinosis universalis and scleroderma, over a period of eight years. The kaleidoscopic course of such cases emphasizes the relationship that most cases of polymyositis bear to the other collagen diseases.

**Group I.**—There were 14 cases in Group I, of which 9 had been diagnosed as progressive
muscular dystrophy and 2 as myasthenia gravis. 3 patients suffered a very acute illness with severe muscular pain and rapid weakness, developed myoglobinuria and died of renal failure. The pathological changes were those of polymyositis and not of idiopathic paroxysmal myoglobinuria. It seems that any condition in which rapid destruction of large amounts of muscle occurs may produce myoglobinuria, which may, in turn, be fatal because of renal damage. However, 9 of the remaining 11 cases had symptoms characteristic of weakness of proximal muscles in the upper and/or lower limbs—symptoms which were often identical with those of muscular dystrophy. Similar muscular weakness was professed by 2 other patients, but the addition of dysphagia, diplopia, fatigability and an undoubted response to Prostigmin resulted in a diagnosis of myasthenia gravis. Undoubtedly some cases in all groups may show a temporary improvement in muscle power after an injection of this drug. 2 patients were children who climbed up their legs and had pseudohypertrophy of the calves, another was a young adult with a picture like limb-girdle dystrophy (Walton and Nattrass, 1954) and 5 fitted the description given by Shy and McEachern (1951) of “menopausal muscular dystrophy". 2 had facial weakness and 2 involvement of the external ocular muscles. None of these cases showed skin changes, constitutional upset or even significant muscle pain and tenderness; in some the course of the disease was slowly progressive, though in others there were features (such as rapid progression, dysphagia and neck weakness, as mentioned by Professor Nattrass) which are not seen in true dystrophy.

Group II.—Of the 12 cases in this group, 7 were initially diagnosed as cases of muscular dystrophy, generally because skin changes or the associated rheumatoid arthritis were transient or unobtrusive. In the children particularly, the skin changes were often limited to the face and hands, and could be nothing more than a tight, shining appearance of the skin; occasionally there were cutaneous ulcers and minimal subcutaneous calcification. Often the skin changes were those of localized scleroderma or acrosclerosis and the associated dysphagia and Raynaud phenomena had been overlooked.

Group III contained 8 cases, either of florid dermatomyositis or of severe rheumatoid arthritis with secondary polymyositis. In passing, it is worth noting that the skin changes could be very variable, looking like exfoliative dermatitis, seborrhoeic dermatitis, lupus erythematosus or scleroderma in different cases.

Group IV.—We have separated the 6 cases in Group IV purely on the basis of an association with malignant disease. 3 were suffering from florid dermatomyositis (like Group III), 1 from severe polymyositis with moderate rheumatoid arthritis (like Group II) and 2 from polymyositis without skin change (like Group I). These last 2 cases correspond closely, from the clinical standpoint, to the condition which Henson, Russell and Wilkinson (1954) and Heathfield and Williams (1954) have called carcinomatous myopathy.

The pathological features are similar in cases of all groups but, as already mentioned, they are not specific, unless combined with the characteristic clinical findings. First one may have variation in fibre size and central migration of sarcolemmal nuclei very like that seen in progressive muscular dystrophy. This occurs particularly in chronic cases or in those which have run a remittent course. Secondly, and most characteristic, there is necrosis and active phagocytosis of muscle fibres, sometimes involving the whole fibre, but more often a segment of it. Thirdly, one usually discovers cellular infiltrates but these may be scanty or even absent (particularly in some Group I or Group IV cases). Finally, there is generally evidence of muscular regeneration; in sections stained with haemalum and eosin or phloxine-methylene blue the regenerating fibres are basophilic, contain greatly enlarged sarcolemmal nuclei with prominent nucleoli, and show a coarse “granularity” in cross section owing to dispersion of the regenerating myofibrils. This coarsely “spotted” appearance is more clearly seen in sections stained with phosphotungstic acid haematoxylin, in which the regenerating fibres are pale.

Prognosis and treatment.—In all groups except the cases with malignant disease, some patients recovered or improved spontaneously, in others the disease became arrested and the patients could lead useful lives, while only a few severe cases died (particularly some in Group III). In all groups, too, some cases responded to ACTH or cortisone. Hence diagnosis from muscular dystrophy is of the greatest importance and often this can only be achieved with certainty by muscle biopsy, though electromyography may help. Our inclusion of all these varied cases under one nosological heading, namely, polymyositis, must not be taken to infer a common aetiology. Although many cases in all groups are clearly related to the other collagen diseases, this is probably not true of all. Our analysis suggests that those cases which show striking cellular infiltrates in the muscle respond best
to cortisone or ACTH; probably in these individuals a hypersensitivity or allergic response is the cause of the disease. In cases without cellular infiltrates, however, which often show no response to similar treatment, it seems possible that the condition may result from some unidentified metabolic or toxic disturbance.

I am indebted to Dr. Raymond D. Adams of the Neurological Service, The Massachusetts General Hospital, Boston, Mass, and to Professor Nattrass for permission to report this material which will be published in detail elsewhere (Walton and Adams, 1956). This work was begun during the tenure of a Nuffield Foundation Fellowship in Neurology; its completion was aided by a grant from the Muscular Dystrophy Association, of America, Inc.

REFERENCES


Dr. P. Bawens:

Variations of the Motor Unit

I should like to discuss some of the significant electroloretical vagaries of the motor unit when pathology visits its distal portion. Some cases make history even in the field of electrodagnosis, and I hope to be forgiven if I outline the one which did so in respect of polymyositis.

Some six years ago, Dr. C. C. Worster-Drought referred to me a typist aged 19, complaining of bilateral weakness of the shoulder-girdles and arms. The condition was said to have started approximately a year earlier with swelling and redness of the palms of the hands, followed by dysphagia and muscle wasting. At the time, the differential diagnosis seemed to lie somewhere between pre-thyrotoxic myopathy and dermatomyositis, with post-scarlatintiform polyneuritis as another suggestion.

Electrodiagnostic investigation revealed that nerve-trunk stimulation in the affected parts produced impaired responses which were well in keeping with the degree of wasting and clinical weakness of the muscles. On direct stimulation of the weak muscles with currents of long duration, sluggish responses were obtained and the intensity-duration curves pointed to partial denervation. As was anticipated from these findings, electromyographic exploration revealed fibrillation at rest, but what was unexpected in the circumstances, and seemed irreconcilable at that time, was the appearance on volition of a motor unit activity pattern which disturbed the whole base line of the cathode ray tube with potentials of low amplitude and short duration—in fact, what might have been expected in a true myopathy, where a large proportion of muscle fibres became inoperative within motor units through primary dysfunction of the muscle fibres.

It seemed here that the impaired activity of a large number of muscle fibres was due to scattered denervation within the motor unit. The phenomenon could be explained by postulating the existence of a pathological process affecting the terminal non-myelinated portions of the ramified neuron causing axon degeneration distally. We labelled this hypothetical condition “distal neuritis” and looked for other cases presenting similar electrodiagnostic features.

In retrospect, it is now moderately certain that the case just outlined was one of “polymyositis” or perhaps more precisely of “neuromyositis”.

Dr. Worster-Drought states that his case proved fatal and that Dr. Peter Daniel of Oxford, on a biopsy specimen, reported that the muscle showed a severe degree of fibrosis with a fairly heavy infiltration of inflammatory cells—mainly lymphocytes but some large mononuclears as well. It appears that the muscle fibres showed a great diversity of size, unusually small fibres predominating and that some of these small fibres had lost their staining reactions and also to some extent their cross striations, while many were broken into short lengths. On the question of intramuscular nerve fibres and endings, the report stated that while a few normal nerve trunks were observed, no nerve endings at the motor end plates were seen.

Since that time, similar cases have been observed, mostly with purely myopathic changes, but sometimes with a neuropathic element so scant that it could not be detected by plotting the intensity-duration curves or testing for nerve conduction and was revealed only on most searching electromyographic exploration by the presence of a few fibrillating muscle fibres.

Frequently, this entity was associated with other pathological processes, as unrelated as carcinoma of the bronchus, steatorrhoea, diabetes and thyrotoxicosis in relation to which it appeared to be secondary. When the primary cause was removed, the electromyographic pattern on volition reverted to an interference pattern compounded of potentials of normal amplitude and duration (Fig. 1).
Fig. 1.—Electromyographic tracings obtained with concentric needle electrode from triceps muscle during progressively increased activity (a) in the normal; (b) in thyrotoxic myopathy showing component potentials of low amplitude and short duration; (c) the same after one month's treatment with iodine and thiouricil; (d) the same two months after thyroidectomy, showing interference pattern compounded of potentials of normal amplitude and duration. Time scale = 10 milliseconds per division.

In this group the readily reversible character of the muscular disturbance suggests the activity of an endogenous toxin acting in the first place as an inhibiting agent, rather than one causing inflammatory or degenerative processes, although I imagine that a more damaging course, resulting in more chronic dysfunction, can ensue in protracted cases. Such a situation prevails even in myasthenia gravis, another reversible condition which, unless checked appears to gravitate progressively towards a permanent impairment of function—tantamount to a secondary myopathy. In the case of a pure polymyositis, the electromyographic tracing on volition is indistinguishable from that of a myopathy whether primary or secondary. It may depart from it where a distal neuritis coexists and can be detected. It does not necessarily follow that the neuritis, when present, is demonstrable. It will be appreciated that where the abolition of the muscle fibre activity precedes the damage to the neuron or neurofibril, it becomes impossible to detect electrodiagnostically the characteristic features of denervation, i.e. sluggish response to direct muscle stimulation with currents of long duration; shift of the intensity-duration curves; or fibrillation on electromyographic exploration. In other words, the myopathic element in polymyositis may mask the neuro-pathic element—the neuritis being detectable electrodiagnostically only if the muscle fibres associated with the degenerated axons are still contractile and excitable.

On a previous occasion, I drew attention to the fact that in chronic myelopathies the discrete action-potentials of large amplitude might be replaced by broader polyphasic ones resulting possibly from a temporal dispersion of the activity of the individual muscle fibres. I went further, and said that this type of disintegrated potential might at times be difficult to distinguish from the true myopathic pattern. Perhaps it is therefore not out of place to reiterate here that, in myelopathies, the small potentials occur repetitively at frequencies approximating those of the normal motor units, while in myelopathies they occur in recognizable discontinuous repetitive trains at those frequencies.

Dr. A. T. Richardson:

Clinical and Electromyographic Aspects of Polymyositis

Polymyositis may be defined as a reaction of striated muscle of unknown etiology of which the essential lesion is muscle fibre necrosis, generally with some evidence of regeneration, accompanied by a variable infiltration of inflammatory cells (Figs. 1A and B). Whether,
in the absence of more exact knowledge of the pathogenesis of this lesion, it can be considered
to indicate a primary inflammatory process comparable to that caused by bacterial, viral or
parasitic invasion of muscle is doubtful, but there is no doubt that it delineates a muscle
disease recognizable not only histologically but often clinically and electrodiagnostically.

In this paper the electromyographic aspects of 20 cases of polymyositis are described with
a brief reference to their clinical features. The series is made up of 13 cases referred for
routine electromyography and 7 selected classical cases of dermatomyositis which were
investigated in addition for abnormalities of neuromuscular transmission.

The 13 cases were referred for electromyography to the Royal Free Hospital and the
Hospital for Sick Children, Great Ormond Street, in the last eighteen months. During this
period of 486 cases referred, in 52 the presence of a myopathic lesion was reported, this
number being made up of the 13 cases of polymyositis, 36 of hereditary muscular dystrophy
and one each of thyrotoxic myopathy, periodic paralysis secondary to potassium-losing
nephritis, and myasthenia gravis. Further analysis of these 52 myopathies, and I am using
the term myopathy in the general sense of a lesion of muscle fibres, shows that in the 21
occurring in adult life the diagnosis of polymyositis was established in 10, while of the 31
occurring in children the incidence was much lower and the diagnosis was established in
only 3.

**CLINICAL DIAGNOSIS OF POLYMYOSITIS**

An early clinical account of polymyositis was given by Steiner (1903) who described it as
"an acute, subacute or chronic disease of unknown origin characterized by gradual onset of
vague and undefined prodromata followed by oedema, dermatitis and multiple muscle
involvement". This description can hardly be bettered except to emphasize that while
acute cases frequently involve the skin (dermatomyositis), the mucous membranes or even
the peripheral nerves (neuromyositis), in the more chronic forms the myopathy may occur
in isolation. Thus apart from the 7 classical cases of dermatomyositis where the diagnosis
was made entirely on the condition of the skin, only 5 of the remaining 13 showed skin
involvement. However, the co-existence of skin erythema and muscle weakness predomin-
antly in the proximal muscles suggested the diagnosis of polymyositis in 3 cases. Of
2 further cases one had calcinosis of the skin and muscles and the second developed sclero-
dermatous changes in the skin thus indicating the true nature of the muscle wasting. The
remaining 8 cases posed the question of the differential diagnosis of muscle wasting and
weakness. In 1 the acute onset of the weakness and wasting with fever, muscle pains and
tenderness immediately suggested the diagnosis of polymyositis and in a further 2 the
association of proximal muscle weakness and dysphagia did so. In 5 cases the diagnosis
of polymyositis on clinical grounds was not made, 3 of these were regarded initially as
cases of motor polyneuritis, 2 were regarded as forms of hereditary muscular dystrophy.

The inclusion in this series of the case showing sclerodermatous changes of the skin seems
justified because the polymyositis dominated the clinical picture, but I have excluded from
this series one case of lupus erythematosus and one of progressive systemic sclerosis in which
muscle involvement, although marked, was overshadowed by the other features of these
diseases. These cases do, however, illustrate the difficulty occasionally encountered in
fixing a diagnostic label to members of the collagen group of diseases showing muscle
involvement. It is noteworthy, however, that in all of 3 cases of periarthritis nodosa with
weakness and wasting that I have examined electromyographically, a pure lower motor
neurone lesion was found and in none could I find evidence of an accompanying myopathy.
Although the presence of Raynaud's phenomenon, raised E.S.R. and increased urinary creatine were an aid to diagnosis, these changes are neither constant nor specific. The association between polymyositis and underlying malignant disease has been emphasized by Denny-Brown (1953) and others. It is, therefore, of interest that 3 of the 13 cases seen in the last eighteen months have developed a carcinoma of the lung and in one, in spite of repeated investigation, five months elapsed after the diagnosis of polymyositis was made before the carcinoma was detected.

**Electrodiagnostic Features of Polymyositis**

Of the total series of 20 cases of polymyositis examined electromyographically 9 showed a purely myopathic lesion and 8 showed the picture which I believe to indicate a combined lower motor neurone and muscle fibre lesion, i.e. a neuromyopathy. At first examination one case of acute polymyositis showed no abnormality but signs of a myopathy developed two months later and 2 of the 7 selected cases of dermatomyositis were within normal limits.

**Criteria of a Myopathic Lesion**

The electrodiagnostic criteria of a myopathic lesion are: brisk responses of muscle to direct stimulation, a high rheobase (10 mA or more) and a normal intensity-duration curve as indicated by 100/1 msec. ratios of less than 2. Electromyographically the findings are no spontaneous activity and on volition an increase in the number of short duration and polyphasic motor unit action potentials which on maximal volition build up to a full interference pattern. This electromyographic change is by no means easily detected, the difficulty being the determination of a significant increase of short duration motor unit potentials. With standard equipment it is particularly difficult, but there are two advances now coming into general use which promise to overcome this difficulty. First, by the use of magnetic tape recording the duration of a number of motor unit potentials sampled from a muscle can be accurately measured. Second, by the use of a frequency analyser an estimation of the number of short duration and polyphasic potentials which are indicated by an increase of the high frequency component (over 300 cycles) can be made (Fig. 2).

![Fig. 2.—Electromyographic tracings and corresponding frequency analysis from: Top—Deltoid muscle in a case of polymyositis. Bottom—Deltoid muscle in partial lower motor neurone lesion. Calibration is 50 cycles at 100 microvolts. Values of the frequency analyser shown below scale in kilocycles per second.](image-url)
Criteria of a Neuromyopathic Lesion

The occurrence of cases of muscle wasting in which the electromyographic finding of short duration motor unit potentials characteristic of a myopathy was combined with the classical signs of lower motor neurone degeneration, i.e. fibrillation potentials and/or change of muscle excitability as demonstrated by intensity-duration curves, has been recognized for some time. Bauwens (1949) originally referred to this condition as distal neuroneitis and postulated a lesion in the lower motor neurone distal to its point of branching. I think, however, there is little doubt that these cases are in fact cases of polymyositis, for not only did 8 of my 20 cases show these features, but since recognizing the association I have been able to demonstrate these findings in any other variety of neuromuscular disease. A somewhat similar electromyographic picture is obtained at the stage of early recovery from lower motor neurone degeneration. However, repeat examination at a later date will allow differentiation of the two conditions by virtue of the fact that recovering lower motor neurone lesions soon exhibit long duration polyphasic potentials (10 msec. or longer) which are never seen in a myopathic lesion.

Neuromuscular Transmission in Polymyositis

The occurrence in cases of polymyositis and dermatomyositis of increasing weakness on exertion suggestive of myasthenia, the apparent response of such muscle weakness to Prostigmin and the demonstration of the pathological changes of myositis in myasthenia gravis (Storlbecker, 1955) prompted an investigation of neuromuscular transmission in polymyositis. Dr. H. C. Churchill-Davidson and I, therefore, measured the response of muscle weakness in 7 selected cases of dermatomyositis, 1 case of lupus erythematosus and 2 cases of acute polymyositis without skin involvement, all of whom complained of excessive fatigue. The method used was measurement of the response of muscles to decamethonium iodide (Churchill-Davidson and Richardson, 1953). In all cases the response was normal, the cases producing the usual depolarizing block and in no instance a block of the competitive inhibition type such as occurs in myasthenia gravis. Similarly we were unable to demonstrate in these cases any convincing response to Prostigmin or Tensilon. It would appear, therefore, that any abnormality of neuromuscular transmission of the myasthenic type in polymyositis, if it occurs at all, is uncommon and these results certainly do not suggest a basis for a diagnostic test.

In summary, therefore, it appears that the diagnosis of polymyositis is readily made on clinical grounds if the muscle wasting is accompanied by the typical skin involvement (dermatomyositis), desquamating erythema, sclerodermatous changes or calcinosis. However, it is the combination of clinical investigations and electromyography that detects this lesion in the majority of cases. Thus the diagnosis can be firmly made if the wasted muscles exhibit the electrodiagnostic criteria of a neuromyopathy and it is strongly suggested if the muscle wasting, shown electrodiagnostically to be a myopathy, occurs in adult life or in the distal muscles or is accompanied by dysphagia or signs of muscle inflammation. There would, however, appear to be a need for more sensitive electromyographic technique to detect myopathic lesions and it is to be hoped that improvements in the method of frequency analysis will supply this.

REFERENCES

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Acknowledgments

I should like to express my gratitude to those physicians at the Royal Free Hospital and Great Ormond Street Hospital who have referred these cases to me and in particular to Dr. G. B. Dowling for permission to investigate the cases of dermatomyositis.